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Paediatric Dermatology Column: Case Report

A case of childhood onset hypopigmented mycosis fungoides initially diagnosed as vitiligo

在兒童期病發的色素減退蕈樣肉芽腫最初被診斷為白癜風病例一宗

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An adolescent boy presented with hypopigmented patches on the trunk and limbs since childhood and was initially diagnosed as vitiligo. Subsequent skin biopsy revealed a hypopigmented variant of mycosis fungoides. The diagnostic pitfalls of hypopigmented mycosis fungoides are discussed and a brief review of the condition is presented.

一名青少年男孩自小童時便在軀幹及四肢有著色素減退的斑塊，最初被診斷爲白癜風。隨後的皮膚活檢揭示爲色素減退性的蕈樣肉芽腫。在此，我們討論色素減退性蕈樣肉芽腫的診斷困難之處及簡要地回顧其臨床表現。

Keywords: Childhood, hypopigmented, mycosis fungoides, vitiligo

關鍵詞：兒童期、色素減退、蕈樣肉芽腫、白癜風

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Case report

A 17-year-old Chinese boy was referred to the dermatology clinic for continuation of care of widespread vitiligo since childhood. The boy had a good past health and unremarkable birth history. He was noted to have multiple asymptomatic hypopigmented patches on the trunk and limbs since early childhood. He was initially seen by the paediatricians with skin biopsy done when he was six years old. The skin biopsy

revealed a marked decrease but not absence of melanocytes and diminished melanin deposition in the dermal-epidermal junction. There was a minimal amount of perivascular lymphocytic infiltrate. The boy had tried topical steroid treatment and supplements and the condition remained static for years. The boy also noticed that the skin hypopigmentation improved with unintentional sun exposure.

Physical examination revealed multiple hypopigmented ill-defined patches on the trunk and limbs (Figures 1 and 2). There were a few small patches with mild erythema and slight textural changes (Figure 3). There were no palpable lymph nodes and no hepatosplenomegaly. A skin biopsy was repeated for lesions on the limbs because of the atypical clinical features. The biopsy revealed scattered atypical lymphoid cells of small to medium size infiltrating the upper dermis and epidermis (Figure 4). The atypical lymphoid cells had irregular, hyperchromatic convoluted nuclei, either in isolation or in small aggregates (Figure 5).

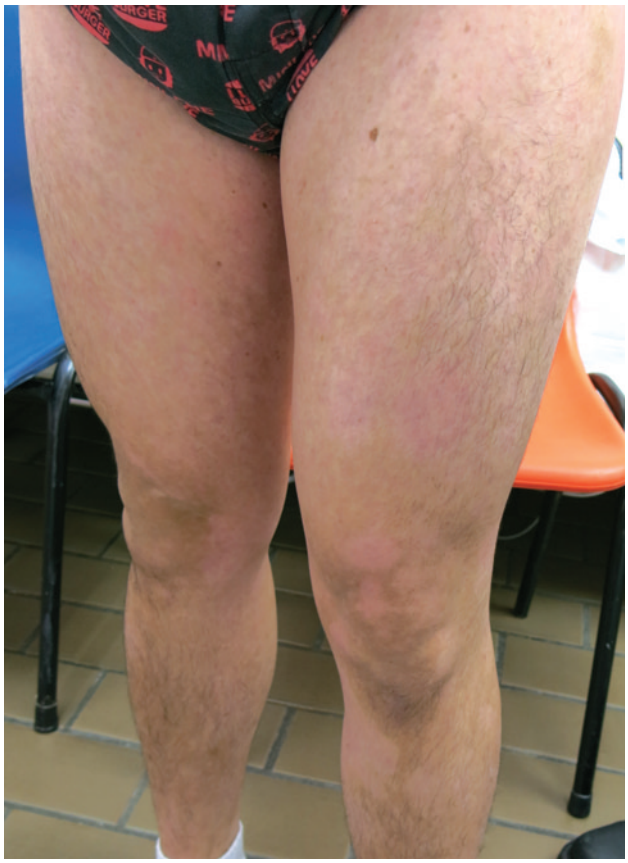

Figure 2. Multiple hypopigmented patches as well as some erythematous patches on the lower limbs.

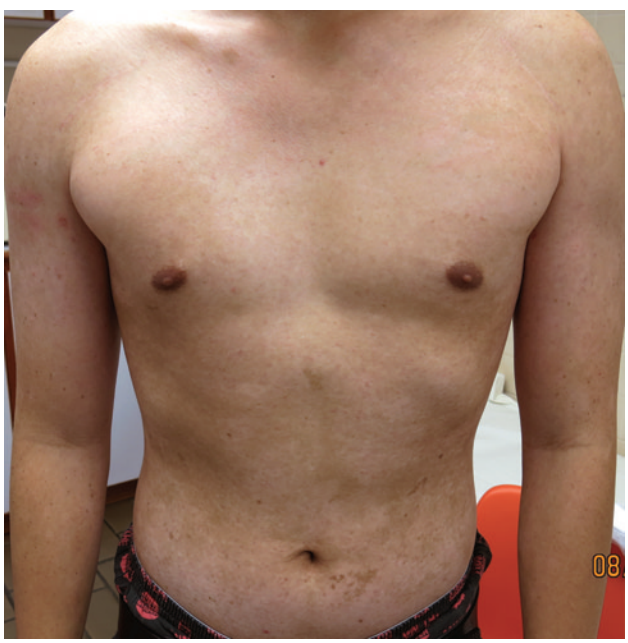

Figure 1. Multiple hypopigmented patches on the trunk and upper limbs.

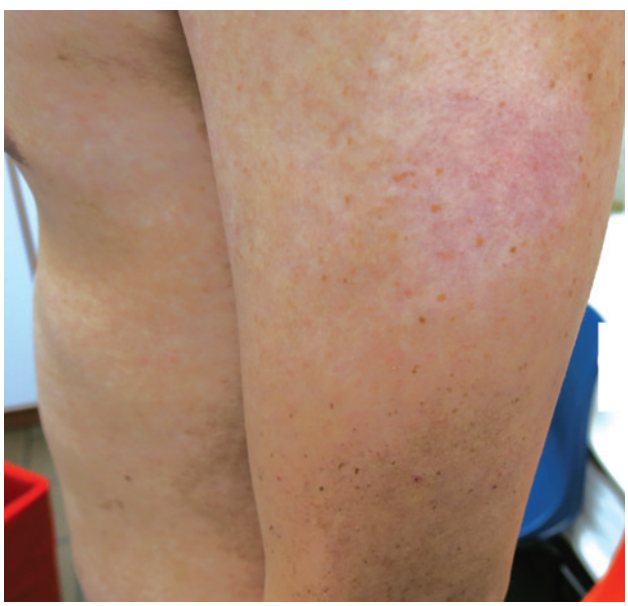

Figure 3. Mild erythematous patch with textural change on the left arm.

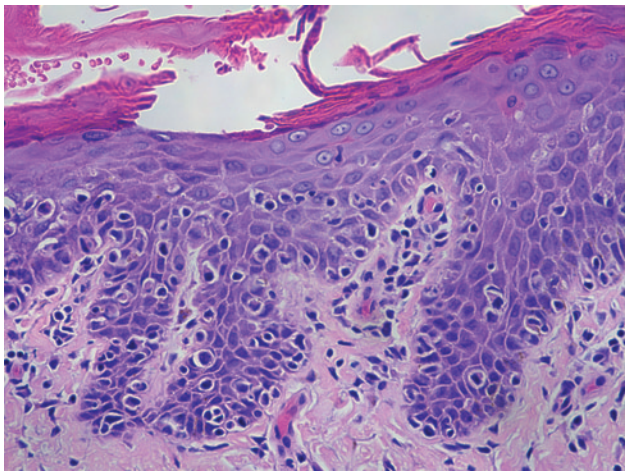

Figure 4. There is epidermotropism, not associated with spongiosis. The infiltrating lymphocytes showed atypia with medium large cerebriform cells and perinuclear halo (H&E, 400X).

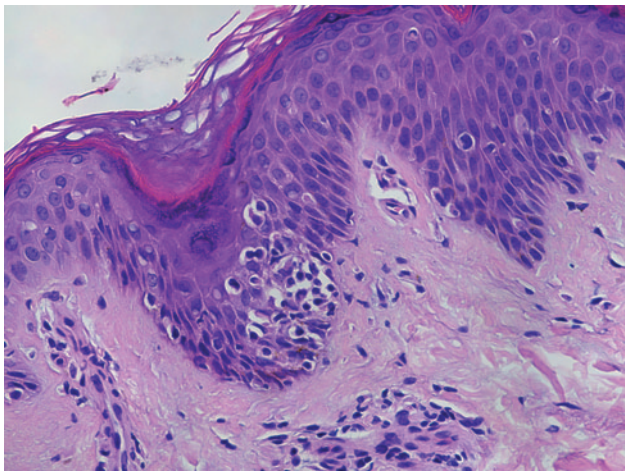

Figure 5. Aggregates of atypical lymphoid cells forming Pautrier's microabscesses (H&E, 400X).

There were minimal or no associated spongiosis and basal melanin was decreased. Immunohistochemical study showed that the atypical lymphoid cells were T cells and positive for CD2, CD3, CD5; CD7 positivity was decreased and the lymphoid cells were predominantly CD8 positive, while CD30 and CD56 were negative. The overall clinical and histopathological features were consistent with the hypopigmented variant of mycosis fungoides (MF).

Discussion

Mycosis fungoides, being the most common type of cutaneous T-cell lymphoma, has been reported to typically occur in older adults.[1] In recent years, there have been increasing reports of childhood-onset MF.[2-4] In a recent paediatric study from Singapore,[5] 46 cases of MF with an age of onset younger than 16 years were found over a period from 2000 to 2008. The mean age of onset of symptoms was 10.3 years with a male predominance. The majority of the children had early stage disease (Stage IA or IB) and all had a favourable clinical outcome with no extracutaneous disease progression during the follow-up period. The histopathological specimens of all cases were reported to have epidermotropism without spongiosis, presence of atypical lymphocytes and individual haloed lymphocytes within the epidermis. These features were all present in our patient.

The hypopigmented variant of MF was reported to be more common in the Asian paediatric population,[2,6] in contrast to Caucasian population, in whom hypopigmented MF is less common.[7] A recent study from Canada, however, found that paediatric hypopigmented MF is not uncommon in North America, although most patients reported were of skin type III to V.[8] Hypopigmented MF is considered a great mimicker and it is important to differentiate this entity from other clinical conditions such as pityriasis lichenoides chronica (PLC), post-inflammatory hypopigmentation secondary to inflammatory dermatoses, pityriasis versicolor and vitiligo. One of the diagnostic challenges is to differentiate hypopigmented MF from PLC. PLC is more common than MF in the paediatric population. However, to complicate the picture, it has been reported to coexist with MF and there are also reports of PLC-like MF.[9] These can only be distinguished by careful histopathological examination of the skin lesions.

Our case was initially diagnosed as vitiligo and in retrospect, our case did show some atypical features at the initial presentation. Firstly, the clinical presentation of widespread of hypopigmented rather than depigmented patches in childhood and the tendency to improve with sun exposure is atypical for vitiligo. Secondly, the histopathology showed diminished but not complete absence of melanocytes as in classic vitiligo; one study found that this feature may be a significant factor to warrant further follow-up and investigation.[10] Thirdly, some of the lesions upon presentation showed atypical clinical features of vitiligo with predominantly hypopigmented patches and some erythematous patches with textural changes. Although the initial skin biopsy did not reveal enough evidence for the diagnosis of MF, the atypical features should alert the clinician for further follow-up of the patient continuously and to repeat skin biopsy as appropriate.

Conclusion

We present a local case of childhood-onset hypopigmented variant of MF. MF in childhood is rare and often not considered in the initial differential diagnosis before the skin biopsy. It is important to consider MF in the differential diagnosis when encountering paediatric patients presenting with hypopigmented skin lesions and atypical clinical features. As hypopigmented MF may be of slow progressive onset in the clinical course, a repeat skin biopsy should be considered even if the initial biopsy did not reveal concrete evidence of MF. There have been no local data in childhood MF, therefore further study in the local epidemiology of this condition is warranted.

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